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Prenatal diagnosis and successful postnatal therapy of an atypical choroid plexus papilloma – Case report and review of literature

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review

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1 (1) Title:

2 Prenatal diagnosis and successfull postnatal therapy of an atypical
3 choroid plexus papilloma – Case report and review of literature

5 (2) Running head:

6 Choroid plexus papilloma - perinatal management

For Peer Review

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3 **Abstract:**
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6 This report shows the perinatal management of a prenatally detected choroid
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8 plexus papilloma (CPP) in an otherwise unaltered singleton pregnancy of an
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10 healthy women. After elective caesarian section a sucessfull embolisation of the
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12 feeding vessel followed by a craniotomy and complete removal of the remaining
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14 tumor was performed. Histological examination confirmed CPP WHO Grade II.
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16
17 One, two and three year follow-up showed a normal development of the girl with a regular
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19 neuro-pediatric status.
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21
22 A review of literature including related papers listed in Pubmed between 1985-2009 was
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24 added to this report.
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28
29 **Key words:**
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31 fetus; CNS; choroid plexus papilloma; ultrasound; embolisation; craniotomy
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3 **1 Introduction:**
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6 2 Choroid plexus papilloma (CPP) is a rare benign intracranial tumor. Affected
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8 3 children typically become conspicuous during their first 24 months showing signs of
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10 4 increased intracranial pressure frequently suffering from headache, nausea and
11
12 5 vomiting, focal neurological impairments and convulsions. The slow growing CPP
13
14 6 may occlude the physiological drainage of the cerebrospinal fluid (CSF), and
15
16 7 additionally secretes CSF itself, provoking a clinical manifestation. Prenatal
17
18 8 detection of a hydrocephalus combined with an intraventricular tumor showing a
19
20 9 hypervascularisation without infiltrative growth pattern is suggestive of a CPP. Due
21
22 10 to the fact that CPP is only representing approximately 3-10% of all pediatric
23
24 11 tumors of the central nervous system (1, 2, 3) large studies do not exist. This
25
26 12 causes a lack of valid recommendations for therapeutical management regarding
27
28 13 CPP and similar benign intracranial tumors of the fetus and the neonate.
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34 14 This case report depicts a feasible perinatal management and successfull postoperative
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36 15 diagnostic and therapeutic regimen in an otherwise healthy child with an ultrasound-detected
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38 16 lesion suspicious for a CPP. A review of literature including related papers listed in Pubmed
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40 17 between 1985-2009 was added to this report.
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Case Report:

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Presentation of a 29 years old women, IG 0P, BMI 26,7, no regular medication, no chronic diseases, no former surgical interventions, with an intact, non-consanguineous singleton pregnancy in the 31st week of gestation: A targeted ultrasound scan (HDI 5000, Phillips Healthcare, Andover, Mass., USA; Voluson 730 Pro, GE Healthcare, Chalfont-St.-Giles, England; 2-5 MHz trans-abdominal probes) revealed an echogenic, slightly perfused mass of about 1 cm in diameter within the posterior horn of the right lateral fetal ventricle and a moderate ipsilateral ventriculomegaly without any additional anomalies (Figure 1-3). There were no signs of invasive growth. Fetal blood sampling revealed normal blood values and a normal female karyotype (46 XX). Serial follow-up scans were scheduled every 14 days and showed a marked increase in tumor size: Within two weeks the lesion having now 6 cm of diameter caused a midline deviation of the cerebral structures and a beginning macrocephaly (Figure 4). Color Doppler and Power Doppler mapping (Figure 5) confirmed increasing vascularization within the tumor with low pulsatility indices. These findings were highly suggestive of a choroid plexus tumor and elective caesarean section without intricacies was performed at completed 35th week of gestation. Confirming the prenatal data the newborn, an otherwise healthy girl, APGAR 7/8/10, length 48 cm, 2230g, UApH: 7.29, had a macrocephaly and a diastasis of the sagittal cranial suture. A cranial MRI and a MR-angiography of the neonate on the first day of life revealed a contrast enhancing tumor in the lateral right ventricle of 8 cm in diameter (Figure 6). The tumor was supplied by the posterior choroid artery approximately 1 mm in diameter. At the sixth day of life an

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3 1 elective conventional angiography confirmed the posterior choroid artery as feeding
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5 2 vessel of the tumor. There were no signs of arterio-venous shunts or additional
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7 3 blood supply by other vessels. Consecutive coiling of the associated artery with 3
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9 4 GDC-Coils and 3 Liquid-Coils lead to a negative blushing of the tumor in the post-
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11 5 procedure angiogram (Figure 7a and 7b). One day after the successful
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13 6 embolization of the feeding vessel a right temporal craniotomy with 3cm
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15 7 corticotomy was performed. The tumor was removed entirely in a complication-free
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17 8 intervention (Figure 8). Only an external ventricular drainage was necessary for 3
18
19 9 days following operation.

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21 10 Histological and immunohistochemical examination revealed a highly differentiated choroid
22
23 11 plexus tumor without nuclear atypia or necrosis and excluded other origins. The tumor
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25 12 showed an increased rate of mitotic activity. Therefore CPP was classified WHO Grade II.
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27 13 One, two and three year follow-up showed a normal neuromotoric development of the child.
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36 **Discussion:**

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38 16 CPPs are benign tumors. CPP originates from the neuroepithelial cells of the
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40 17 choroid plexus. The plexus is located in the body and inferior horn of the lateral
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42 18 ventricles, in the interventricular foramen and in the third and fourth ventricle. CPPs
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44 19 can therefore be found in all these locations described above. In pediatric patients
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46 20 CPPs are found in the lateral ventricle with 71% of all cases (2, 4, 5). CPP growth
47
48 21 in the third and fourth ventricle are rare (6, 7, 8). In adults the allocation of CPP is
49
50 22 slightly different: main region of initial growth of CPPs is the fourth ventricle (76%)
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52 23 as it was shown in a single center population with a median age of 36 years (9).
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1 Extraventricular occurrence of CPPs is an exception (10). Bilateral ventricular
2 growth has been regularly reported (11, 12). CPPs are described as tumors
3 showing a non-invasive and slow growth (12) but due to their specific localization
4 they are accidentally occluding the liquor drainage and causing an internal
5 hydrocephalus. The situation may be aggravated by the potential ability of a CPP
6 producing cerebrospinal fluid (CSF) itself.

7 In our case we saw an unusually rapid tumor expansion from 1 to 8 cm in
8 approximately 6 weeks.

9 Table 1 shows previous studies regarding CPP.

10
11 In adults CPPs account for less than 1% of all brain tumors (13). In contrast 3-10%
12 of all pediatric tumors of the central nervous system are choroid plexus papillomas
13 (1, 2, 3). The median age of children at time of diagnosis is 22.5 months. One-half
14 of the patients are reported to be younger than 2 years when CPP becomes
15 evident (4). This is one of the rare cases where CPP was suspected prenatally.

16
17 CPP is a polygenetic disorder (14). Different genetic loci have been described to be
18 possibly connected to the development of this benign tumor. Known associations
19 are mutations of the TP53 tumor suppression gene (17p13.1) shown in germline
20 cells of an adult with CPP and also osteosarkoma (15). However, dysregulation of
21 protein p53 is far more often present in cases with choroid plexus carcinoma than
22 in CPPs (16). Alterations in the hSNF5/INI1 gene (22q11.2) considered important
23 in choroid plexus carcinomas do not seem to have this impact on CPP (17). The

1 following alterations have been reported in CPP (18): +7q (65%), +5q (62%), +7p
2 (59%), +5p (56%), +9p (50%), +9q (41%), +12p, +12q (38%), +8q (35%), -10q
3 (56%), -10p, and -22q (47%). CPPs in childhood show different chromosomal
4 alterations with +8q, +14q, +12, and +20q suggesting a different genetic pathway.
5 A duplication of 9p has been shown in few cases of choroid plexus carcinoma and
6 CPP as well.

7 In our case no chromosomal imbalances could be detected.

8 There are two syndromes known which are considered to coincide with CPP:

9 Aicardi-Goutières-Syndrome (AGS) (12) and Hypomelanosis of Ito (HI) (14). Both
10 syndromes are associated with other significant problems. Patients with AGS, a
11 multigenetic disorder with similarities to characteristic features of congenital
12 infections, have malformations of the brain, like agenesis of corpus callosum and a
13 considerable developmental impairment (12, 19, 20). HI is described as an
14 accumulation of neurological and skeletal anomalies. Its name originates from the
15 characteristic small hypopigmented macules on the skin along the lines of
16 Blaschko of the affected individuals. In combination with the chromosomal
17 translocation X;17(q12;p13) HI has been found with CPP (21, 22).

18 The child in our case did not show any signs of these specific features.

19 Another factor in the development of a CPP may be seen in intrauterine infections.

20 There is evidence of similarities between DNA found in choroid plexus tumors and
21 ependymomas and those of simian virus 40 (23). Secondary artificial choroid
22 plexus tumor growth as been induced experimenally by adding typical viral product,
23 T antigen, in transgenic mice (24).

1 Two case-reports implicating connections between the presentation of Hippel-
2 Lindau-disease, a genetic disorder located on chromosome 3 p25/26 and
3 characterised by haemangiomas and/or haemangioblastomas situated in the brain
4 stem, cerebellum, spinal cord or retina. CPP or invasive choroid plexus tumor
5 respectively having been seen coexisting in the same individual with Hippel-Lindau
6 disease in adulthood (25, 26).

7
8 CPP-cells are similar to choroid-plexus-cells which are cuboidal epithelial cells
9 based on papillary fibrovascular tissue. This explains why CPP-cells are able to
10 produce CSF (11). CPP normally consists of highly differentiated cells without
11 signs of increased mitotic activity, nuclear atypia, necrotic areas and/or altered
12 vascularisation. Tumors consisting of these cells are classified as CPP WHO grade
13 I. Minor calcification of the fibrovascular stroma is not an uncommon sign.

14 Expansion of cysts accompanying a CPP may be seen (27), deterioration of
15 cerebral tissue through cystic development is found sporadically (28). In our
16 discussed case we saw a higher mitotic activity, leading to a classification
17 analogous to WHO grade II. Due to the higher grading in our case the tumor has to
18 be labelled *atypical* CPP. Choroid plexus carcinomas usually classify as WHO
19 grade III.

20 CPP can show both an aneuploid or, like in our case, a normal karyotype (29).

21 Malignant transformation of CPPs is a rare (30).

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3 1 The most common clinical signs of children with CPP are provoked by raised
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6 2 intracranial pressure caused by increasing hydrocephalus (6). Children may suffer
7
8 3 from headaches, nausea and vomiting, focal neurological deficits, macrocephaly
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10 4 and epileptic seizures (5). Hydrocephalus is evident in approximately 85-87% of all
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12 5 cases (4, 31). There seems to be no prognostic impact in occurrence of
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14 6 hydrocephalus. It correlates poorly with the actual size and pathological features of
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16 7 the tumor (4).
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22 9 Diagnostic procedures/imaging of CPP are normally performed after first symptoms
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24 10 have been noted in an affected individual. MRI displays CPP as a lobulated
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26 11 contrast-enhancing lesion similar to grey matter on T1w images often accompanied
27
28 12 by an enlarged lateral ventricle. T2w imaging presents a hyperintense lesion (1,
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30 13 32). Computed tomography shows an isodense to hyperdense lesion often
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32 14 presenting intra-tumoral calcifications with uniform contrast enhancement (6, 28,
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34 15 30). CPP in transabdominal ultrasound imaging of an affected fetus shows an
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36 16 echogenic, lobulated *intraventricular tumor* typically associated with *feeding*
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38 17 *vessel(s) in color flow Doppler* mapping. Adding the sonographic presentation of
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40 18 *hydrocephalus* the *triad* postulated for CPP by Adra et al.,1994, (6) is completed.
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44 19 In our case we report on an echogenic intraventricular tumor with ipsilateral
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46 20 ventriculomegaly (Figure 1-3). This tumor showed an significant tumor growth
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48 21 causing a midline deviation of the cerebral structures and a macrocephaly as well
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50 22 (Figure 4). At the end of pregnancy nearly the complete ipsilateral cerebral
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52 23 hemisphere was displaced by the tumor. Color Doppler and Power Doppler
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1 mapping (Figure 5) confirmed increasing vascularization within the tumor with low
2 pulsatility indices. In some areas of this lobulated tumor we also could detect minor
3 calcifications.

4 We were able to confirm the triad postulated by Adra et al., 1994.

5
6 Complete surgical excision of the tumor tissue is considered to be the treatment of
7 choice for CPPs (4, 9). Surgical intervention is complicated by a risk of
8 intraoperative hemorrhage which is a common and often lethal emergency in
9 resection of CPPs (4, 11). In about 15% of CPPs being operated a gross total
10 resection (GTR) could not be achieved due to vascular problems (5, 9). Significant
11 blood-loss may be prevented by preoperative embolisation (33). Even partial
12 embolisation may decrease the severity of intraoperative hemorrhage.

13 In 24-41% of reported cases a ventriculo-peritoneal shunt was necessary (4, 31).

14 Preoperative symptoms have proved to be an independent marker for the need of
15 ventriculo-peritoneal shunting after surgery took place (4). In our case the patient
16 needed an external ventricular drainage for 3 days after the intervention.

17 Postoperative brain collapse causing subdural fluid collections may occur and
18 require a subduro-peritoneal shunting drainage in 8-14% of cases (4, 31).

19 Therefore preoperative controlled drainage of the affected ventricle and
20 intraoperative anchorage adjacent to the cortical incision are recommended by
21 some authors (4). In our case we did not seal the temporal corticotomy by using a
22 biological glue. After the procedure moderate subdural hygroma developed
23 postoperatively and resolved spontaneously.

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3 1 In 22% of cases temporary swallowing dysfunction has been described as an
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5 2 notable postoperative complication with frequent need for tracheostomy or
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7 3 percutan gastrostomy (9).
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10 4 Adding chemo- and/or radiation-therapy does not significantly influence the overall
11
12 5 survival rate. Adequate therapy in case of recurrence of symptoms where a subtotal
13
14 6 resection (STR) of the tumor had been performed seems to be the re-operation (5),
15
16 7 which is reported to be necessary in about 50% of such cases (9). Radiation may
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18 8 be an alternative option in the rare constellation of STR after recurrence of a CPP
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20 9 (9) and in cases where a surgical intervention is not feasible (34).
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27 11 CNS neoplasms are responsible for 5-20% of deaths in the fetal or neonatal period
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29 12 (1). The prognosis of CPP is favourable for patients treated with GTR. Malignant
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31 13 transformation is described in pediatric patients with STR but seems to be a very
32
33 14 rare incident (5, 30, 35). The overall 5-year survival rate in cases with GTR is
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35 15 reported with 100% (4). In our case the outcome of the otherwise healthy girl was
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37 16 good. Complete neuro-pediatric status was performed and no signs of neurological
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39 17 impairment could be found. Physical development was also regular. Follow-up was
40
41 18 three years.
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48 **Conclusion:**

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50 21 This is the first reported case of an atypical CPP being diagnosed prenatally and
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52 22 successfully embolised and resected postnatally with a favourable outcome of the
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54 23 affected child. This report shows that it is feasible to detect tumors like CPP
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1 prenatally via ultrasound scans. A prenatal detection may lead to a proper
2 counselling of the parents and perinatal management improving the short- and
3 intermediate-term outcome of children with CPP or similar benign intracranial
4 tumors.

For Peer Review

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1 (1) Title:

2 Prenatal diagnosis and successfull postnatal therapy of an atypical
3 choroid plexus papilloma – Case report and review of literature

5 (2) Running head:

6 Choroid plexus papilloma - perinatal management

For Peer Review

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1 **Abstract:**

2 This report shows the perinatal management of a prenatally detected choroid
3 plexus papilloma (CPP) in an otherwise unaltered singleton pregnancy of an
4 healthy women. After elective caesarian section a sucessfull embolisation of the
5 feeding vessel followed by a craniotomy and complete removal of the remaining
6 tumor was performed. Histological examination confirmed CPP WHO Grade II.

7 One, two and three year follow-up showed a normal development of the girl with a regular
8 neuro-pediatric status.

9 A review of literature including related papers listed in Pubmed between 1985-2009 was
10 added to this report.

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12 **Key words:**

13 fetus; CNS; choroid plexus papilloma; ultrasound; embolisation; craniotomy

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3 **1 Introduction:**
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6 2 Choroid plexus papilloma (CPP) is a rare benign intracranial tumor. Affected
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8 3 children typically become conspicuous during their first 24 months showing signs of
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10 4 increased intracranial pressure frequently suffering from headache, nausea and
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12 5 vomiting, focal neurological impairments and convulsions. The slow growing CPP
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14 6 may occlude the physiological drainage of the cerebrospinal fluid (CSF), and
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16 7 additionally secretes CSF itself, provoking a clinical manifestation. Prenatal
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18 8 detection of a hydrocephalus combined with an intraventricular tumor showing a
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20 9 hypervascularisation without infiltrative growth pattern is suggestive of a CPP. Due
21
22 10 to the fact that CPP is only representing approximately 3-10% of all pediatric
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24 11 tumors of the central nervous system (1, 2, 3) large studies do not exist. This
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26 12 causes a lack of valid recommendations for therapeutical management regarding
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28 13 CPP and similar benign intracranial tumors of the fetus and the neonate.
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34 14 This case report depicts a feasible perinatal management and successfull postoperative
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36 15 diagnostic and therapeutic regimen in an otherwise healthy child with an ultrasound-detected
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38 16 lesion suspicious for a CPP. A review of literature including related papers listed in Pubmed
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40 17 between 1985-2009 was added to this report.
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Case Report:

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Presentation of a 29 years old women, IG 0P, BMI 26,7, no regular medication, no chronic diseases, no former surgical interventions, with an intact, non-consanguineous singleton pregnancy in the 31st week of gestation: A targeted ultrasound scan (HDI 5000, Phillips Healthcare, Andover, Mass., USA; Voluson 730 Pro, GE Healthcare, Chalfont-St.-Giles, England; 2-5 MHz trans-abdominal probes) revealed an echogenic, slightly perfused mass of about 1 cm in diameter within the posterior horn of the right lateral fetal ventricle and a moderate ipsilateral ventriculomegaly without any additional anomalies (Figure 1-3). There were no signs of invasive growth. Fetal blood sampling revealed normal blood values and a normal female karyotype (46 XX). Serial follow-up scans **were scheduled every 14 days and** showed a marked increase in tumor size: Within two weeks the lesion having now 6 cm of diameter caused a midline deviation of the cerebral structures and a beginning macrocephaly (Figure 4). Color Doppler and Power Doppler mapping (Figure 5) confirmed increasing vascularization within the tumor with low pulsatility indices. These findings were highly suggestive of a choroid plexus tumor and elective caesarean section without intricacies was performed at completed 35th week of gestation. Confirming the prenatal data the newborn, an otherwise healthy girl, APGAR 7/8/10, length 48 cm, 2230g, UApH: 7.29, had a macrocephaly and a diastasis of the sagittal cranial suture. A cranial MRI and a MR-angiography of the neonate on the first day of life revealed a contrast enhancing tumor in the lateral right ventricle of 8 cm in diameter (Figure 6). The tumor was supplied by the posterior choroid artery approximately 1 mm in diameter. At the sixth day of life an

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3 1 elective conventional angiography confirmed the posterior choroid artery as feeding
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5 2 vessel of the tumor. There were no signs of arterio-venous shunts or additional
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7 3 blood supply by other vessels. Consecutive coiling of the associated artery with 3
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9 4 GDC-Coils and 3 Liquid-Coils lead to a negative blushing of the tumor in the post-
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11 5 procedure angiogram (Figure 7a and 7b). One day after the successful
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13 6 embolization of the feeding vessel a right temporal craniotomy with 3cm
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15 7 corticotomy was performed. The tumor was removed entirely in a complication-free
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17 8 intervention (Figure 8). Only an external ventricular drainage was necessary for 3
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19 9 days following operation.

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25 10 **Histological and immunohistochemical examination revealed a highly differentiated choroid**
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27 11 **plexus tumor without nuclear atypia or necrosis and excluded other origins.** The tumor
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29 12 showed an increased rate of mitotic activity. Therefore CPP was classified WHO Grade II.
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31 13 One, two and three year follow-up showed a normal neuromotoric development of the child.
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36 15 **Discussion:**

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39 16 CPPs are benign tumors. CPP originates from the neuroepithelial cells of the
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41 17 choroid plexus. The plexus is located in the body and inferior horn of the lateral
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43 18 ventricles, in the interventricular foramen and in the third and fourth ventricle. CPPs
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45 19 can therefore be found in all these locations described above. In pediatric patients
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47 20 CPPs are found in the lateral ventricle with 71% of all cases (2, 4, 5). CPP growth
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49 21 in the third and fourth ventricle are rare (6, 7, 8). In adults the allocation of CPP is
50
51 22 slightly different: main region of initial growth of CPPs is the fourth ventricle (76%)
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53 23 as it was shown in a single center population with a median age of 36 years (9).
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1 Extraventricular occurrence of CPPs is an exception (10). Bilateral ventricular
2 growth has been regularly reported (11, 12). CPPs are described as tumors
3 showing a non-invasive and slow growth (12) but due to their specific localization
4 they are accidentally occluding the liquor drainage and causing an internal
5 hydrocephalus. The situation may be aggravated by the potential ability of a CPP
6 producing cerebrospinal fluid (CSF) itself.

7 In our case we saw an unusually rapid tumor expansion from 1 to 8 cm in
8 approximately 6 weeks.

9 Table 1 shows previous studies regarding CPP.

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11 In adults CPPs account for less than 1% of all brain tumors (13). In contrast 3-10%
12 of all pediatric tumors of the central nervous system are choroid plexus papillomas
13 (1, 2, 3). The median age of children at time of diagnosis is 22.5 months. One-half
14 of the patients are reported to be younger than 2 years when CPP becomes
15 evident (4). This is one of the rare cases where CPP was suspected prenatally.

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17 CPP is a polygenetic disorder (14). Different genetic loci have been described to be
18 possibly connected to the development of this benign tumor. Known associations
19 are mutations of the TP53 tumor suppression gene (17p13.1) shown in germline
20 cells of an adult with CPP and also osteosarkoma (15). However, dysregulation of
21 protein p53 is far more often present in cases with choroid plexus carcinoma than
22 in CPPs (16). Alterations in the hSNF5/INI1 gene (22q11.2) considered important
23 in choroid plexus carcinomas do not seem to have this impact on CPP (17). The

1 following alterations have been reported in CPP (18): +7q (65%), +5q (62%), +7p
2 (59%), +5p (56%), +9p (50%), +9q (41%), +12p, +12q (38%), +8q (35%), -10q
3 (56%), -10p, and -22q (47%). CPPs in childhood show different chromosomal
4 alterations with +8q, +14q, +12, and +20q suggesting a different genetic pathway.
5 A duplication of 9p has been shown in few cases of choroid plexus carcinoma and
6 CPP as well.

7 In our case no chromosomal imbalances could be detected.

8 There are two syndromes known which are considered to coincide with CPP:

9 Aicardi-Goutières-Syndrome (AGS) (12) and Hypomelanosis of Ito (HI) (14). Both
10 syndromes are associated with other significant problems. Patients with AGS, a
11 multigenetic disorder with similarities to characteristic features of congenital
12 infections, have malformations of the brain, like agenesis of corpus callosum and a
13 considerable developmental impairment (12, 19, 20). HI is described as an
14 accumulation of neurological and skeletal anomalies. Its name originates from the
15 characteristic small hypopigmented macules on the skin along the lines of
16 Blaschko of the affected individuals. In combination with the chromosomal
17 translocation X;17(q12;p13) HI has been found with CPP (21, 22).

18 The child in our case did not show any signs of these specific features.

19 Another factor in the development of a CPP may be seen in intrauterine infections.

20 There is evidence of similarities between DNA found in choroid plexus tumors and
21 ependymomas and those of simian virus 40 (23). Secondary artificial choroid
22 plexus tumor growth as been induced experimenally by adding typical viral product,
23 T antigen, in transgenic mice (24).

1 Two case-reports implicating connections between the presentation of Hippel-
2 Lindau-disease, a genetic disorder located on chromosome 3 p25/26 and
3 characterised by haemangiomas and/or haemangioblastomas situated in the brain
4 stem, cerebellum, spinal cord or retina. CPP or invasive choroid plexus tumor
5 respectively having been seen coexisting in the same individual with Hippel-Lindau
6 disease in adulthood (25, 26).

7
8 CPP-cells are similar to choroid-plexus-cells which are cuboidal epithelial cells
9 based on papillary fibrovascular tissue. This explains why CPP-cells are able to
10 produce CSF (11). CPP normally consists of highly differentiated cells without
11 signs of increased mitotic activity, nuclear atypia, necrotic areas and/or altered
12 vascularisation. Tumors consisting of these cells are classified as CPP WHO grade
13 I. Minor calcification of the fibrovascular stroma is not an uncommon sign.

14 Expansion of cysts accompanying a CPP may be seen (27), deterioration of
15 cerebral tissue through cystic development is found sporadically (28). In our
16 discussed case we saw a higher mitotic activity, leading to a classification
17 analogous to WHO grade II. Due to the higher grading in our case the tumor has to
18 be labelled *atypical* CPP. Choroid plexus carcinomas usually classify as WHO
19 grade III.

20 CPP can show both an aneuploid or, like in our case, a normal karyotype (29).

21 Malignant transformation of CPPs is a rare (30).

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1 The most common clinical signs of children with CPP are provoked by raised
2 intracranial pressure caused by increasing hydrocephalus (6). Children may suffer
3 from headaches, nausea and vomiting, focal neurological deficits, macrocephaly
4 and epileptic seizures (5). Hydrocephalus is evident in approximately 85-87% of all
5 cases (4, 31). There seems to be no prognostic impact in occurrence of
6 hydrocephalus. It correlates poorly with the actual size and pathological features of
7 the tumor (4).

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9 Diagnostic procedures/imaging of CPP are normally performed after first symptoms
10 have been noted in an affected individual. MRI displays CPP as a lobulated
11 contrast-enhancing lesion similar to grey matter on T1w images often accompanied
12 by an enlarged lateral ventricle. T2w imaging presents a hyperintense lesion (1,
13 32). Computed tomography shows an isodense to hyperdense lesion often
14 presenting intra-tumoral calcifications with uniform contrast enhancement (6, 28,
15 30). CPP in transabdominal ultrasound imaging of an affected fetus shows an
16 echogenic, lobulated *intraventricular tumor* typically associated with *feeding*
17 *vessel(s) in color flow Doppler* mapping. Adding the sonographic presentation of
18 *hydrocephalus* the *triad* postulated for CPP by Adra et al.,1994, (6) is completed.

19 **In our case we report on an echogenic intraventricular tumor with ipsilateral**
20 **ventriculomegaly (Figure 1-3). This tumor showed an significant tumor growth**
21 **causing a midline deviation of the cerebral structures and a macrocephaly as well**
22 **(Figure 4). At the end of pregnancy nearly the complete ipsilateral cerebral**
23 **hemisphere was displaced by the tumor. Color Doppler and Power Doppler**

1 mapping (Figure 5) confirmed increasing vascularization within the tumor with low
2 pulsatility indices. In some areas of this lobulated tumor we also could detect minor
3 calcifications.

4 We were able to confirm the triad postulated by Adra et al., 1994.

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6 Complete surgical excision of the tumor tissue is considered to be the treatment of
7 choice for CPPs (4, 9). Surgical intervention is complicated by a risk of
8 intraoperative hemorrhage which is a common and often lethal emergency in
9 resection of CPPs (4, 11). In about 15% of CPPs being operated a gross total
10 resection (GTR) could not be achieved due to vascular problems (5, 9). Significant
11 blood-loss may be prevented by preoperative embolisation (33). Even partial
12 embolisation may decrease the severity of intraoperative hemorrhage.
13 In 24-41% of reported cases a ventriculo-peritoneal shunt was necessary (4, 31).
14 Preoperative symptoms have proved to be an independent marker for the need of
15 ventriculo-peritoneal shunting after surgery took place (4). In our case the patient
16 needed an external ventricular drainage for 3 days after the intervention.
17 Postoperative brain collapse causing subdural fluid collections may occur and
18 require a subduro-peritoneal shunting drainage in 8-14% of cases (4, 31).
19 Therefore preoperative controlled drainage of the affected ventricle and
20 intraoperative anchorage adjacent to the cortical incision are recommended by
21 some authors (4). In our case we did not seal the temporal corticotomy by using a
22 biological glue. After the procedure moderate subdural hygroma developed
23 postoperatively and resolved spontaneously.

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3 1 In 22% of cases temporary swallowing dysfunction has been described as an
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6 2 notable postoperative complication with frequent need for tracheostomy or
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8 3 percutan gastrostomy (9).
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10 4 Adding chemo- and/or radiation-therapy does not significantly influence the overall
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12 5 survival rate. Adequate therapy in case of recurrence of symptoms where a subtotal
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14 6 resection (STR) of the tumor had been performed seems to be the re-operation (5),
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16 7 which is reported to be necessary in about 50% of such cases (9). Radiation may
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18 8 be an alternative option in the rare constellation of STR after recurrence of a CPP
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20 9 (9) and in cases where a surgical intervention is not feasible (34).
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27 11 CNS neoplasms are responsible for 5-20% of deaths in the fetal or neonatal period
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29 12 (1). The prognosis of CPP is favourable for patients treated with GTR. Malignant
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31 13 transformation is described in pediatric patients with STR but seems to be a very
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33 14 rare incident (5, 30, 35). The overall 5-year survival rate in cases with GTR is
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35 15 reported with 100% (4). In our case the outcome of the otherwise healthy girl was
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37 16 good. Complete neuro-pediatric status was performed and no signs of neurological
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39 17 impairment could be found. Physical development was also regular. Follow-up was
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41 18 three years.
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48 20 **Conclusion:**

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50 21 This is the first reported case of an atypical CPP being diagnosed prenatally and
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52 22 successfully embolised and resected postnatally with a favourable outcome of the
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54 23 affected child. This report shows that it is feasible to detect tumors like CPP
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3 1 prenatally via ultrasound scans. A prenatal detection may lead to a proper
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5 2 counselling of the parents and perinatal management improving the short- and
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7 3 intermediate-term outcome of children with CPP or similar benign intracranial
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9 4 tumors.
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Table 1: Diagnosis and outcome of CPP in former investigations

Study	Sex	No. of cases	Localisation	Sonographic presentation: Adra's Triad (6)	time of diagnosis	Intervention	Outcome
Adra et al., 1994	female	1	third ventricle	Confirmed triad	33rd week of gestation	no intervention	SPP and neonatal death
Buckle & Smith, 2007	male	1	third ventricle	not sufficiently specified: Confirmed ventriculomegaly and intraventricular tumor	2 months	not specified	not specified
Frye et al., 2007	female	1	right lateral ventricle	no ventriculomegaly	2 months; additionally revealing Aicardi-Syndrome	craniotomy and GTR at the age of 7 years due to symptomatic issues	clinical status (e.g. sleep disturbances) unchanged
Fujimura et al., 2004	male	1	right and left lateral ventricle	Confirmed triad	10 months	repeated left and right craniotomy with STR	significant reduction of excessive CSF-production
Noguchi et al., 2004	not specified	1	third ventricle	not sufficiently specified: Confirmed ventriculomegaly and intraventricular tumor	40th week of gestation	frontal craniotomy on day 15 with GTR without shunting	7 days post interventionem: subdural effusions on treated side
Otten et al., 2006	male	1	right lateral ventricle	Confirmed triad	4 months	embolisation and sequential parietal craniotomy with GTR	good neurological outcome at 4-month follow-up
Patanakar et al., 2000	male	1	right lateral ventricle	Confirmed triad	30 months	no intervention	exitus letalis
Stafrace &	not	1	right	not sufficiently specified:	1 months	embolisation	not specified

Molloy, 2008	specified		cerebellopontine angle	Confirmed ventriculomegaly and intraventricular tumor		and resection	
Chow et al., 1999	1 female/ 3 male	4	4/4 lateral ventricle	not sufficiently specified: Confirmed ventriculomegaly and intraventricular tumor	19 months (11-200 months)	STR 2/4 later GTR 1/2, 2nd STR and Chemotherapy 1/2; Initial GTR 2/4	OS (54-98 months): 100%. Moderate/severe delayed neurological outcome
McEvoy et al., 2000	no sex difference reported	25	not specified; mixed CPP/CPC-population	not sufficiently specified: Confirmed ventriculomegaly and intraventricular tumor	17 months (1-138 months); 11/25 cases > 12 months	GTR in 25/25 cases	in 3/25 second look surgery. 1/25 postoperative death. Median survival 76 months
Pencalet et al., 1998	not specified for CPP	25	not specified for CPP	not sufficiently specified: Confirmed ventriculomegaly in most cases and intraventricular tumor	not specified for CPP	GTR 24/25; STR 1/25	OS at 5 years 100%
Krishnan et al., 2004	22 female/ 19 male	41	6/41 right lateral ventricle; 1/41 left lateral ventricle; 31/41 fourth ventricle; 3/41 third ventricle	Hydrocephalus in 34% of cases. Intraventricular tumor in 100% of cases. No color-flow mapping described	36 years (6 months-74 years)	GTR 23/41; STR in 18/41. Postoperative radiation 4/41; 1/41 radio-chemo-therapy; 1/41 gamma-knife	OS at 5 years: GTR 100%; STR 94%.

CPP: Choroid Plexus Papilloma; CPC: Choroid Plexus Carcinoma; GTR: Gross total resection; STR: Subtotal Resection; OS: Overall survival; SPP: spontaneous vaginal delivery

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3 Fig. 1: Illustration of choroid plexus papilloma within the posterior horn of the
4 right lateral ventricle combined with ipsilateral ventriculomegaly
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6 Fig. 2 (30+3 gw): Trans-abdominal ultrasound-scan, transverse view,
7 showing an echogenic tumor in the right lateral ventricle (arrows)
8

9 Fig. 3 (31+6 gw): Trans-abdominal ultrasound-scan, transverse view,
10 showing tumor-growth in the lateral ventricle and beginning midline-deviation
11

12 Fig. 4 (34+6 gw): Trans-abdominal ultrasound-scan, transverse view,
13 showing ongoing tumor-growth in the lateral ventricle
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15 Fig. 5 (34+6 gw): Trans-abdominal ultrasound-scan, transverse view,
16 showing power-doppler of the feeding vessel (arrow) of a tumor in the right ventricle
17

18 Fig. 6 (1st day postnatally): cMRI T1w showing an
19 hyperintense tumor in the posterior horn of the right lateral ventricle (arrows)
20

21 Fig. 7a (6 days postnatally): embolisation of the feeding
22 vessel (arrow): pre-interventional angiogram
23

24 Fig. 7b (6 days postnatally): successfull embolisation of the feeding
25 vessel (arrow): post-interventional angiogram
26

27 Fig. 8 (8th day postnatally): cMRI T1w after craniotomy and surgical removal of
28 the tumor in the right lateral ventricle (circle). Subdural hemorrhage (arrows)
29 and frontal pneumatic residues (quadrats)
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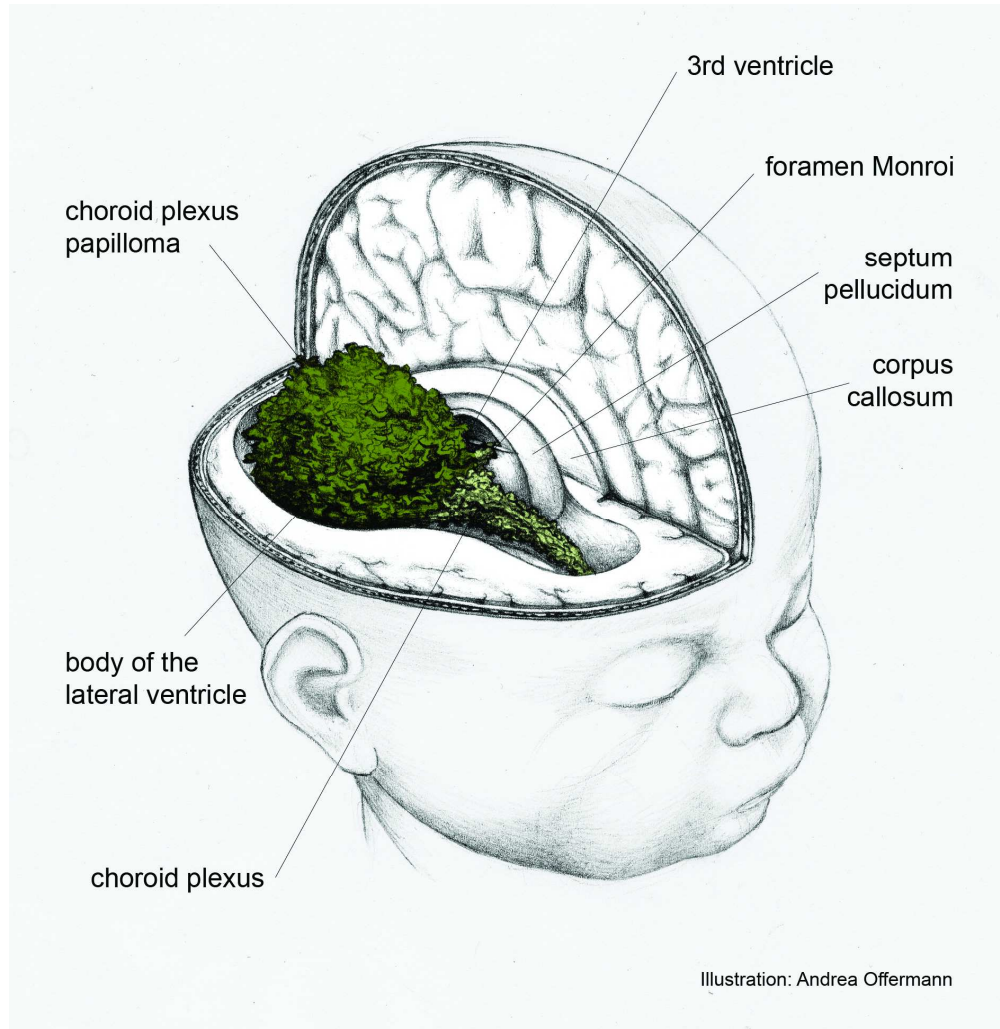


Fig. 1: Illustration of choroid plexus papilloma within the posterior horn of the right lateral ventricle combined with ipsilateral ventriculomegaly
160x164mm (300 x 300 DPI)

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Fig. 2 (30+3 gw): Trans-abdominal ultrasound-scan, transverse view, showing an echogenic tumor in the right lateral ventricle (arrows)

52x39mm (300 x 300 DPI)

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Fig. 3 (31+6 gw): Trans-abdominal ultrasound-scan, transverse view, showing tumor-growth in the lateral ventricle and beginning midline-deviation

45x39mm (300 x 300 DPI)



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Fig. 4 (34+6 gw): Trans-abdominal ultrasound-scan, transverse view, showing ongoing tumor-growth in the lateral ventricle

52x39mm (300 x 300 DPI)

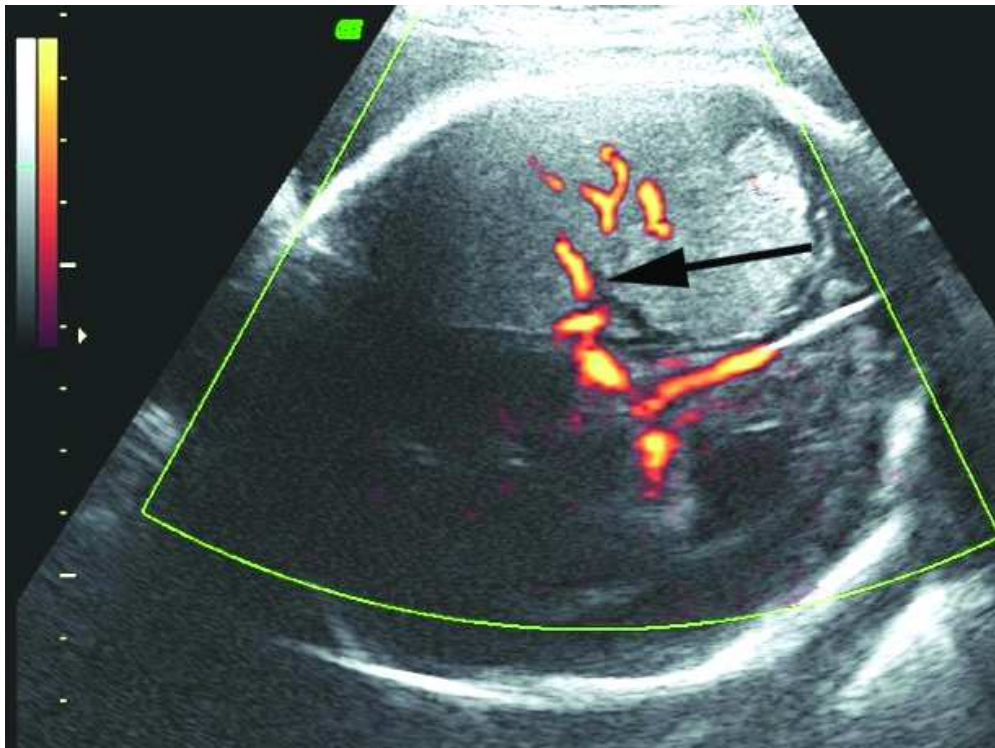


Fig. 5 (34+6 gw): Trans-abdominal ultrasound-scan, transverse view, showing power-doppler of the feeding vessel (arrow) of a tumor in the right ventricle

51x38mm (300 x 300 DPI)

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Fig. 6 (1st day postnatally): cMRI T1w showing an hyperintense tumor in the posterior horn of the right lateral ventricle (arrows)

212x282mm (23 x 23 DPI)

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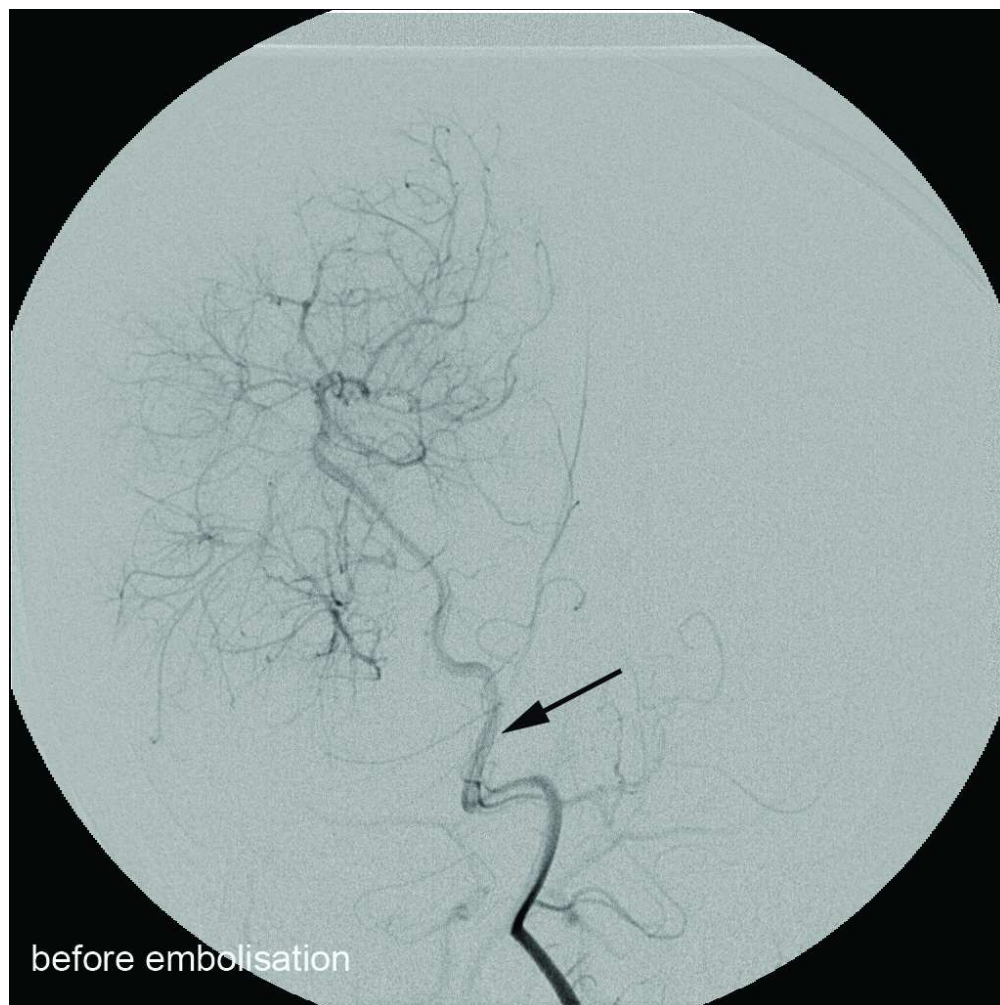


Fig. 7a (6 days postnatally): embolisation of the feeding vessel (arrow): pre-interventional angiogram 86x86mm (300 x 300 DPI)

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Fig. 7b (6 days postnatally): successful embolisation of the feeding vessel (arrow): post-interventional angiogram
1130x1130mm (23 x 23 DPI)



Fig. 8 (8th day postnatally): cMRI T1w after craniotomy and surgical removal of the tumor in the right lateral ventricle (circle). Subdural hemorrhage (arrows) and frontal pneumatic residues (quadrats)

212x282mm (23 x 23 DPI)